

Hypoparathyroidism and Difficult to Control Hypocalcemia

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Introduction

- Autoimmune polyglandular syndrome type 1 (APS-1) is an autosomal recessive disorder characterized by autoimmune multi-organ involvement.
- Clinical manifestations are widely variable though classic triad is composed by mucocutaneous candidiasis, hypoparathyroidism and adrenal failure.
- Treatment is based on supplementation of the various deficiencies (1).

Case Presentation

- An 18-year-old female diagnosed at age 12 with APS-1 (Nail dystrophy & candida infection at age 7, primary hypoparathyroidism at age 9 and Addison's disease at age 10).
- The main challenge in her management was maintaining of normocalcemia with avoidance of hypercalciuria and nephrocalcinosis.
- She had several admissions with symptomatic hypocalcemia, (adjusted calcium as low as 1.6mmol/l) and required intravenous calcium infusions. On one occasion the infusion was complicated by skin necrosis.
- Normocalcemia was rarely achieved despite being on calcium carbonate (1200mg three times as day) and vitamin D analogue (Figure 1).

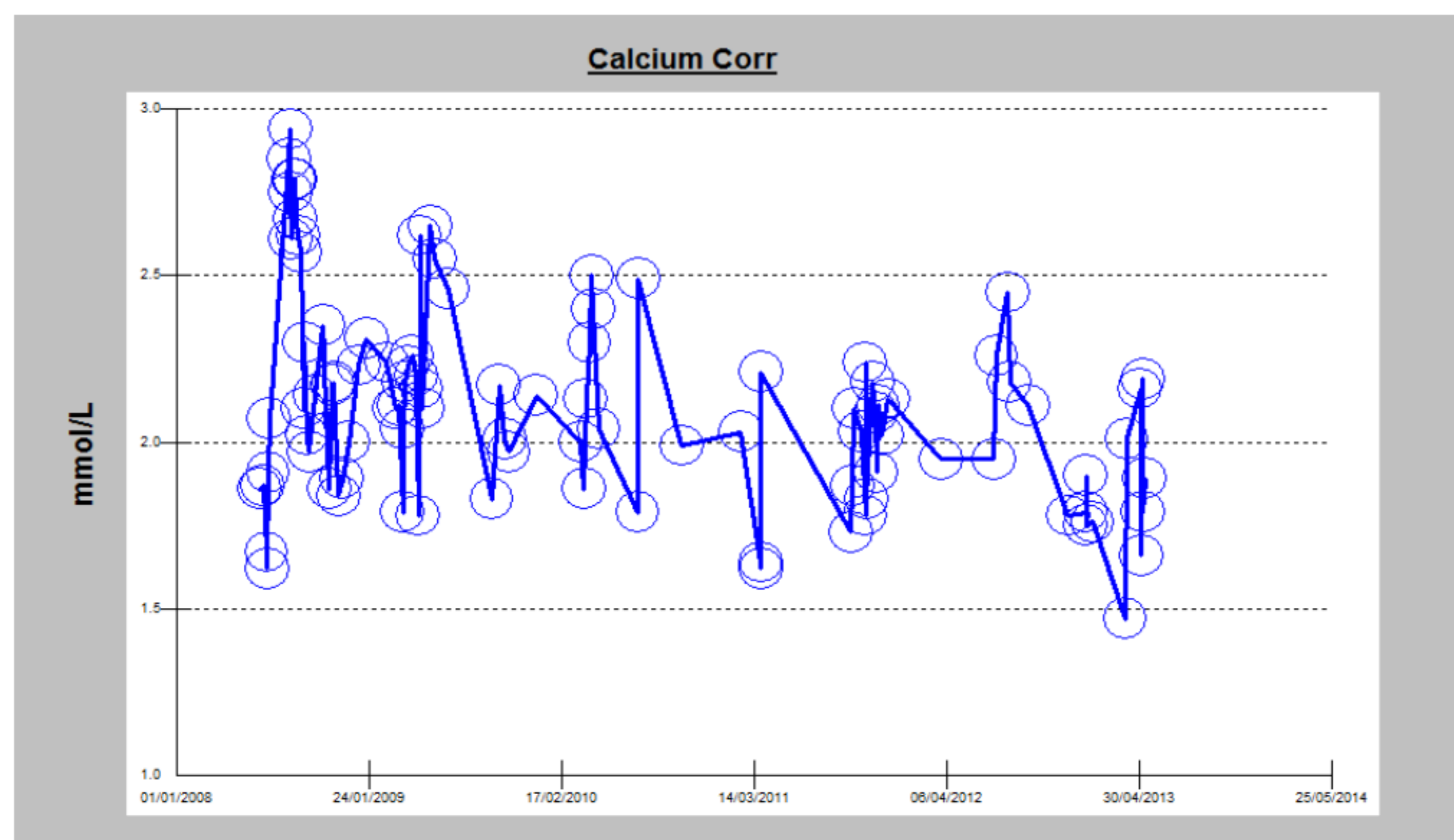


Figure 1

- In 2014 she was started on recombinant human PTH (rPTH). Initial dose was 20 mcg /per day gradually titrated to three times a day in combination with oral calcitriol and calcium.
- Fluctuations of calcemia did not improve. Aiming to improve quality of life, rPTH was delivered by a pump 0.3units /hr (Figure 2).



Figure 2

- Initially, near normal calcium (1.9-2.2 mmol/l) was achieved. Later, patient was admitted with abdominal pain and vomiting and was found to have adjusted calcium level of 3.03mmol/l.
- During admission calcium fluctuated between 2.4-2.95 mmol/l despite suspending calcium and vitamin D analogue supplements as well as adjustment of rPTH infusion rate (Figure 3).

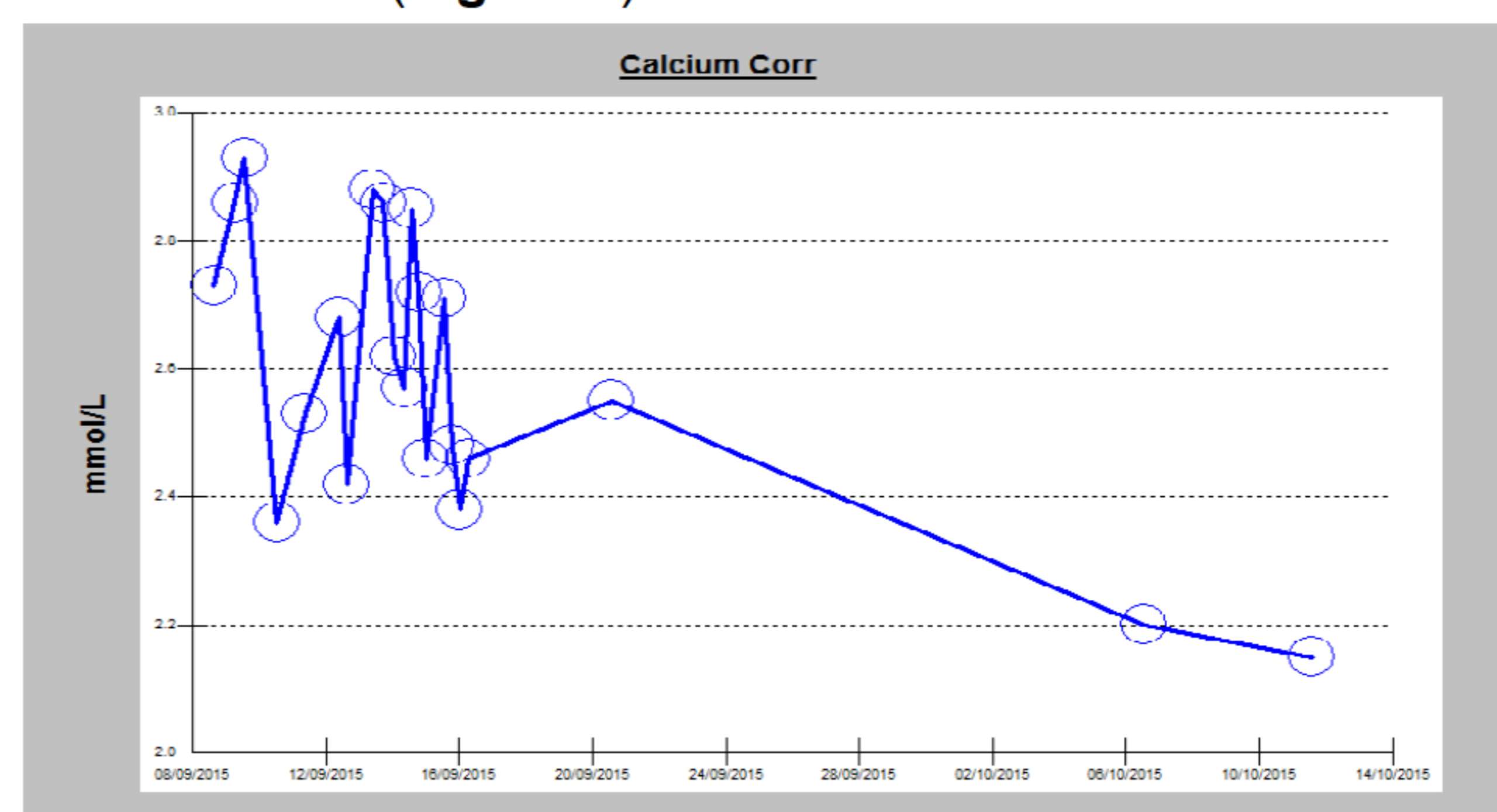


Figure 3

- It later transpired that the patient was giving her self boluses of PTH to avoid symptoms of hypocalcemia.
- Once he mother took charge of pump device, calcium normalised at 2.38mmol/l and she was discharged on rPTH rate 0.25 unit/hr and with calcium 600 mg twice daily and cholecalciferol 1000 units daily.
- At 2 weeks follow up , corrected calcium level was 2.2 and 2.15 mmol/l (Figure 3)

Discussion

- Normocalcemia was difficult to maintain in this patient using conventional therapy with calcium and vitamin D analogue.
- Several studies have reported that normocalcaemia can be better achieved and maintained with the use of rhPTH.
- A study by the National Institutes of Health (NIH) in US showed an almost normalization of the diurnal rhythm of PTH, calcium and phosphate levels in response to rhPTH(1-34) therapy delivered by an infusion pump (2).
- In our case, calcium level improved initially while on infusion pump. The latter fluctuation in her calcium level mandated close observation of her calcium level especially during her admissions with hypercalcemia.
- Subsequently, her hypercalcemia was found to be intentional and due to the inappropriate self administration of rPTH boluses to avoid the feared risk of hypocalcemia induced seizure that she had experienced in the past. However, a secondary gain can not be excluded.

References

- 1- George S. Eisenbarth, M.D., Ph.D., and Peter A. Gottlieb, M.D, Autoimmune Polyendocrine Syndromes, N Engl J Med 2004;350:2068-79
2. Jens Bollerslev^{1,2,*}, Lars Rejnmark^{3,*}, Claudio Marcocci⁴, Dolores M Shoback⁵, Antonio Sitges-Serra⁶, Wim van Biesen⁷ and Olaf M Dekkers⁸, European Society of Endocrinology Clinical Guideline: Treatment of chronic hypoparathyroidism in adults, European Journal of Endocrinology (2015) 173, G1–G20

